

Wunderlich Syndrome as the First Manifestation of Renal Cell Carcinoma

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INTRODUCTION

Renal cell carcinoma (RCC) classically presents as a triad of hematuria, loin pain, and a palpable mass. However, RCCs are increasingly found as incidental findings on the radiological imaging ordered for other reasons.⁽¹⁾ The spontaneous rupture of the kidney, or Wunderlich syndrome, is very rarely the first presentation of RCC. We describe a clinical case of spontaneous renal hemorrhage as the first manifestation of RCC that was treated with therapeutic embolization and early radical nephrectomy.

CASE REPORT

A 63-year-old woman presented to the emergency department with severe generalized abdominal pain. She had marked pallor and was triaged to the resuscitation area. She described a history of 12-hour severe, constant pain with sudden onset and no associated radiation, which became progressively worse. There had been no precipitating factors or history of trauma. She did not have a previous medical disorder and did not take any medications. She was a non-smoker.

On examination, her vital signs were normal. Examination elicited generalized guarding and tenderness over the left flank. Her bowel

sounds were present and a poorly defined mass was palpable over the left side of her abdomen. When she was asked for a urine specimen, a gross hematuria was noted. Serial blood tests showed a drop in serum hemoglobin from 12.5 g/dL to 10.1 g/dL after just one hour, with a concomitant decrease in hematocrit level (from 37% to 29%). She was immediately resuscitated with intravenous fluids and cross matched with 4 units of blood. Then, a computed tomography (CT) scan was performed, which revealed a 14-cm mass involving the left kidney consistent with bleeding complicating a probable RCC at the lower pole of the kidney (Figures 1 and 2). The patient's hemoglobin levels continued to decrease, and she was transfused with 4 units of blood. Her creatinine level was normal.

The patient underwent emergency embolization (Figures 3 and 4). Then, she was admitted to the Intensive Care Unit for close observation and strict bed-rest. Moreover, a broad-spectrum antibiotic was commenced. She also had a central line inserted for access. Her hemoglobin level stabilized.

Five days after presentation, the patient underwent nephrectomy. Surgical excision was difficult, but

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Figure 1. Coronal CT reconstruction demonstrating irregularity of the renal contour with extensive hemorrhage into the perirenal fat. Areas of calcification are demonstrated within a lower pole mass consistent with a renal cell carcinoma.



Figure 2. Axial CT image showing extensive retroperitoneal hemorrhage. Mixed attenuation within the hemorrhage reflects the temporal heterogeneity of the bleeding. The epicenter of the hemorrhage appears to be a calcified soft tissue density mass consistent with a renal cell carcinoma.



Figure 3. Selected image from Digital Subtraction Angiography demonstrating paucity of vessels at the lower pole with contrast blush (as shown by arrow) indicating intraparenchymal hemorrhage.



Figure 4. Selected post-embolization image demonstrating absence of contrast blush with no flow of contrast to the lower pole of the kidney.

with careful dissection, the kidney and associated hematoma were removed, and the specimen was delivered. Later, the patient made a remarkable recovery and was discharged after 2 weeks. Histological examination revealed an extensively necrotic and disrupted RCC that could not be nuclear graded. The adrenal gland and the renal vein margin were free of tumor consistent with a pT2 stage (Figures 5 and 6). Within the 6-month follow-up, there was no sign of recurrence on repeated CT and her creatinine levels remained normal.

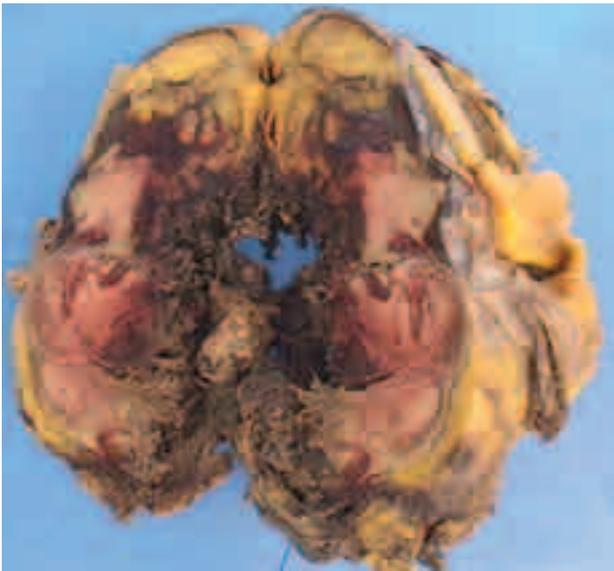


Figure 5. Resected kidney specimen image showing rupture of the lower pole and features of infarction and necrosis consistent with hemorrhage and embolization.

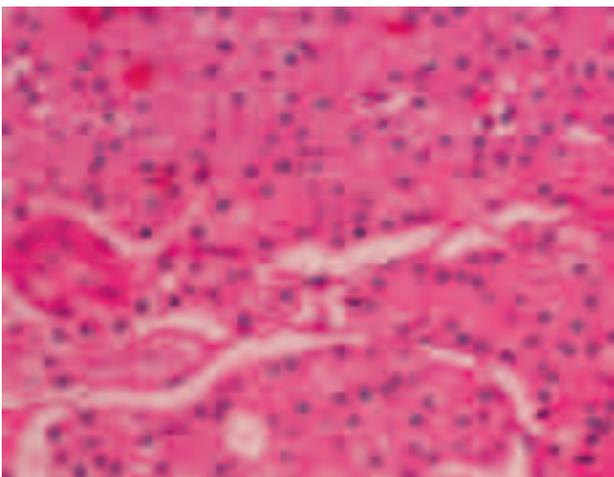


Figure 6. High power view of specimen showing conventional clear cell carcinoma.

DISCUSSION

With increasingly sophisticated imaging techniques, the classic presentation of loin pain, palpable mass, and frank hematuria are less commonly seen. Instead, RCCs are now more commonly discovered as ‘incidentalomas’ on the radiological imaging for alternative abdominal pathologies.⁽¹⁾ The spontaneous rupture of the kidney, however, is rarely the first clinical presentation of RCC.

In 1856, Carl Reinhold August Wunderlich described the spontaneous bleeding of the kidney with dissection of blood into the subcapsular and/or perinephric spaces.⁽²⁾ Wunderlich syndrome is uncommon and is usually caused by a benign disease.⁽³⁾ The most recent meta-analysis by Zhang and colleagues in 2002 reviewed 165 patients with spontaneous perirenal hemorrhage due to various causes between 1985 and 1999.⁽⁴⁾ Seventy percent of subjects with hemorrhage were due to benign causes, including vascular disease, infection, and neoplasia. Overall, neoplastic causes accounted for 61.2% of these cases, with benign and malignant causes approximately divided equally (Table).

In treating Wunderlich syndrome, some urologists favor an early or immediate exploratory surgery.⁽⁵⁾ Others prefer interventional radiology to stop an acutely hemorrhaging vessel.⁽⁶⁾ There are logical reasons to avoid surgery in the acute scenario when the patient’s general condition are not fully stabilized, but embolization with the intention of delayed surgery also results in a more difficult resection due to adherence. There are currently no guidelines favoring either approach.

The efficiency of CT to diagnose renal tumors at the time of bleeding is a further area of concern. Computed tomography remains one of the most

Etiology of spontaneous renal hemorrhage*

Etiology	Percentage (%)
Tumor	61.2
Benign	31.5
Malignant	29.7
Vascular Disease	17.0
Infection	2.42
Miscellaneous	12.7
Idiopathic Hemorrhage	6.7

*Adapted from Zhang and colleagues⁽⁴⁾ with their permission.

reliable modalities in diagnosing retroperitoneal hemorrhage and RCCs.⁽⁷⁾ Kendall and colleagues, however, found that 60% of subjects showed a RCC undiagnosed at the time of initial CT⁽⁸⁾, which is in agreement with Zhang's meta-analysis where CT performed at the time of hemorrhage was only partially efficient at identifying renal tumors (sensitivity 0.57).⁽⁴⁾ For malignant tumors diagnosed on initial CT, radical nephrectomy is eventually required. However, in the renal hemorrhages thought to be benign, embolization may be the sole modality used. If these patients were later found to have a malignancy on the follow-up CT, delayed surgery would not only affect the overall resectability of the tumor, but also the clinical staging of the disease.^(9,10)

Wunderlich syndrome is a rare phenomenon that usually occurs due to a benign renal pathology, but a significant proportion, as in this case, are associated with malignancy. Underlying malignancy may be missed on initial CT scan. One must, therefore, be suspicious when confronted with any spontaneous perirenal hemorrhage. The cornerstones of management include resuscitation, embolization, and nephrectomy, but timing, as well as the treatment modality, is important. Where RCC is diagnosed on initial CT, immediate embolization and early resection, when the patient is stable, is a safe and reasonable approach to the timely management of this difficult condition.

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CONFLICT OF INTEREST

None declared.

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